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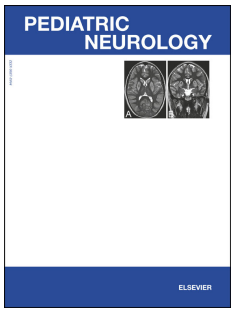


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# Alternating IV nerve palsy and ptosis as a first sign of childhood ocular myasthenia gravis

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The authors have not published or submitted any related papers about this case.

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A previously healthy five-year-old girl was referred due to diplopia of sudden onset. The patient presented with a moderate *right* ptosis, left inferior oblique overaction and superior oblique underaction, and a positive Bielschowsky head tilt test to the left. This suggested a *left* IV nerve palsy but the right ptosis remained undefined. (Fig.1A) Ophthalmic and neurological examination and cranial MRI were otherwise normal. 10 days later, a *right* IV nerve palsy with a *right* ptosis and left adduction deficit was evident. One month after initial presentation, a *left* ptosis and *left* IV nerve palsy were present. Simpson test was positive after 70sec and the ptosis improved after the ice pack test. Repetitive nerve stimulation of the facial nerve demonstrated an abnormal decr ement. Serum acetylcholine receptor (AChR) antibody level was elevated. The diagnosis of ocular myasthenia gravis (OMG) was confirmed. A chest MRI excluded a thymoma. Treatment with pyridostigmine was initiated with a good response after an increase to 60 mg a day. Fourth nerve palsy resolved completely after six months and no generalization was observed.

OMG can mimic oculomotor disorders and should be considered in children with unusual and alternating oculomotor symptoms. Differential diagnoses are neoplasm, trauma, inflammatory or vascular causes. Ophthalmic signs such as ptosis or ophthalmoplegia present in about 90% of children with myasthenia with or without generalization.[1] In prepubertal children ocular symptoms appear mostly at disease onset and spontaneous remission is common. The risk for generalization decreases after two years of disease duration[2] but the development of generalized symptoms is still possible after a period of three years[3]. Untreated pediatric patients with OMG may have a relatively low risk of developing generalized symptoms[2]. Thymectomy, when performed before the onset of generalized symptoms, showed a trend toward protection from the development of generalized symptoms but no prospective data

are available.[2] AChR antibody level, although not always positive, repetitive nerve stimulation and, most sensitive (if the first remain negative) stimulated single fiber EMG is recommended to confirm the diagnosis. Pyridostigmine is the first line treatment of pediatric OMG. Treatment with steroids in young children is ambivalent due to the side effects and may not be necessary in pediatric OMG. Immuno-suppressive agents may reduce the conversion of adult OMG to generalized myasthenia gravis but seems not to influence the course of the disease in children.[2, 4]

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**Figure:**

Presentations with alternating sides of ptosis and 4$^{th}$ nerve palsy.


1st presentation
2nd presentation
3rd presentation